Maryland
DEPARTMENT OF HEALTH

# Spinraza (Nusinersen)

Spinraza is a survival motor neuron-2 (SMN2)-directed antisense oligonucleotide indicated for the treatment of spinal muscular atrophy (SMA) in pediatric and adult patients.*

## I. Criteria for Initial Approval

Spinraza will be considered for coverage when **ALL** of the criteria below are met, confirmed with supporting medical documentation.

- Diagnosis of SMA Type I, II, or III; AND
- Diagnosis by a neurologist with expertise in the diagnosis of SMA; AND
- Genetic testing confirming both:
    - 5q SMA homozygous gene deletion, homozygous gene mutation, or compound heterozygous mutation;
    - At least 2 copies of SMN2; AND
- Patient is not dependent on invasive ventilation or tracheostomy; AND
- Patient is not dependent on non-invasive ventilation beyond use for naps and nighttime sleep; AND
- Patients with Type II and III SMA must have some functional upper extremity use; AND
- Prescribed by a neurologist experienced in treating SMA; AND
- Baseline motor examination completed utilizing at least one of the following exams (based on patient age and motor ability) to establish baseline motor ability:
    - Hammersmith Infant Neurological Exam (HINE); or
    - Hammersmith Functional Motor Scale Expanded (HFMSE); or
    - Upper Limb Module Test (non-ambulatory); or
    - Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP-INTEND)

## II. Criteria for Continuation of Therapy

All of the criteria for initial therapy in Section I. must be met**; AND**

- Repeat motor testing completed since the most recent Spinraza dose (and not more than 1 month prior to the next scheduled dose) using the same motor test done to establish baseline motor ability, unless it is determined that the original test is no longer appropriate; AND
- Repeat motor testing must document a response to treatment as defined by the following:

- HINE:
  - Improvement or maintenance of previous improvement of at least 2 points (or max score of 4) in ability to kick (improvement in at least 2 milestones); OR
  - Improvement or maintenance of previous improvement of at least 1 point increase in motor milestones of head control, rolling, sitting, crawling, standing or walking (consistent with improvement by at least 1 milestone); AND
  - Improvement or maintenance of previous improvement in more HINE motor milestones than worsening;

- HFMSE:
  - Improvement or maintenance of improvement of at least a 3 point increase in score;
- ULM:
  - Improvement or maintenance of previous improvement of at least 2 point increase in score;

- CHOP-INTEND:
  - Improvement or maintenance of previous improvement of at least 4 point increase in score.

## III. Dosing/Administration

Spinraza must be administered according to the current FDA labeling guidelines for dosage and timing. Spinraza must be administered intrathecally by a physician or other healthcare professional experienced in performing lumbar punctures.

- The recommended dosage is 12 mg (5 mL) per administration
- Initiate Spinraza treatment with 4 loading doses; the first three loading doses should be administered at 14-day intervals; the 4th loading dose should be administered 30 days after the 3rd dose
- A maintenance dose should be administered once every 4 months thereafter

## IV. Length of Authorization

Spinraza will initially be preauthorized for 4 loading doses when criteria are met. Preauthorization is valid for 90 days. Each Spinraza maintenance dose (continuing therapy) must be preauthorized.

## V. Billing Code/Information

HCPCS code: J2326 –Injection, nusinersen, 0.1 mg; 1 billable unit = 0.1 mg

*Prior authorization of benefits is not the practice of medicine nor the substitute for the independent medical judgment of a treating medical provider. The materials provided are a component used to assist in making coverage decisions and administering benefits. Prior authorization does not constitute a contract or guarantee regarding member eligibility or payment. Prior authorization criteria are established based on a collaborative effort using input from the current medical literature and based on evidence available at the time.*

**Approved by MDH Clinical Criteria Committee: 7/1/2017**
**Last Reviewed Date: 1/4/2021**

Revision History:

*1/4/2021- Effective 1/1/2021, MCOs are required to cover Spinraza per program guidance. Prior clinical criteria statements referencing MCO Carve Out are no longer applicable and therefore removed.*